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NERVOUS SYMPTOMS IN CASES OF CONGENITAL SYPHILIS.

BY

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NERVOUS SYMPTOMS IN CASES OF CONGENITAL SYPHILIS.

Some years ago I published a pamphlet (reprinted, with slight alterations, from the "St. Andrew's Medical Graduates' Transactions," vol. iv., 1868) on Nervous Affections in Inherited Syphilis. At that time I had had but one post-mortem examination. The patient was a girl, the daughter of the patient Joseph Mx., whose case is the second of two related by me in this Journal for July, 1874. She had epileptic fits. She died of typhoid fever, and no lesion of a syphilitic nature was discovered post-mortem. Thus I learned nothing from this case. In the "Brit. Med. Journal,"* May 18, 1872, I have reported a case of hemiplegia, previously unpublished, in a woman aged 22, who was manifestly the subject of inherited syphilis. That patient was in good general health, and is, I hope, still living.

In the "London Hospital Reports," vol. i., 1864, I gave particulars of several cases in a syphilitic family. There were four children. The eldest, a girl of 18, had good teeth, but remains of old iritis, and scars of ulcerations at the angles of the mouth. Her general health seemed good. A girl of 15, whose sight was much impaired from choroiditis, and who had imperfect hemiplegia. This child had the malformation in the teeth (*Vide infra*). A girl, aged 12, who had the same malformation and choroiditis. A boy aged 8, paraplegic, partly idiotic, and who had had several fits. This boy was quite blind; both optic discs were found to be dirty white, margins obscure, vessels small, and fundus hazy.

I have, unfortunately, no post-mortem evidence as to the pathology of the nervous system in any case of congenital syphilis. In the case I am about to relate I obtained no autopsy. The subject, however, is of such vast importance that I draw attention to some of the clinical facts of it, with the hope of making investigations more methodical.

Now and then I see children and young people who present unmistakeable signs of hereditary syphilis, and who have also definite nervous symptoms, *e. g.*, hemiplegia, epileptiform

* There are, unfortunately, two printer's blunders in the article in that Journal. In the 17th line from the top, for "such evidence of hemiplegia," read "such evidence of congenital syphilis;" and in line 23 from the bottom for "his upper central" read "her upper central."

convulsions, &c. I speak only of cases in which the patient's own body presents decisive signs of congenital syphilis. But in the absence of post-mortem examinations on such cases, it would be not sagacity but laziness to assume confidently that there was a connection—to speak of them as, for example, “hemiplegia *caused by* congenital syphilis.” I had recently under my care a child who was hemiplegic, and who had remains of double choroiditis; another young girl who had a large hole, no doubt the result of syphilitic disease, in her palate, and hemiplegia. The hemiplegia in both was of the common kind, affecting the face, arm, and leg of one side. That these patients were syphilitic was clear enough. But what medical man does not see cases of hemiplegia of this sort in children, where there is no trace of probability of syphilitic taint? To speak of “eliminating all other causes” is not really so practical as it sounds, for nothing is more difficult than to form a reasonable conjecture even as to the cause of hemiplegia in childhood (excluding epileptic hemiplegia with double optic neuritis, cases with valvular disease, and other easy cases). And I wish to point out that even assuming that we could by some means be quite confident that congenital syphilis caused a child's hemiplegia, we should be only on the threshold of a very difficult pathological inquiry. The following is necessarily speculative, as it is not founded on post-mortem examinations, but it states correctly what takes place in a certain form of hemiplegia from acquired syphilis in adults.

As to the general *position* of the changes causing the hemiplegia in children, there is no room for doubt. The hemiplegia is (in range) just like that so constantly met with in the hemiplegia of persons of middle age from local softening, and from cerebral hæmorrhage. The difficulty is the *pathology*. We may say there is (in long standing cases of hemiplegia of children) atrophy of the opposite side of the brain; this would be highly probable. We might go further and say that in all probability there was in such old standing cases a cyst in the corpus striatum. But it would be a very crude statement to say that syphilis caused atrophy of the brain and a cyst. In order to tell whether syphilis brought these changes about we want more facts as to the state of the brain at the onset of the hemiplegia. The atrophy and the local cyst are relics of long past acute changes. I suppose the cyst represents past softening from thrombosis, and the atrophy is a consequence of that local damage. If, then, we

assume as a provisional hypothesis that there was at first local softening from thrombosis, the next question is—"By what process did the artery become occluded?" Having regard to what is well known of one common mode of causation of hemiplegia by syphilis in adults, the question is, does congenital syphilis lead to hemiplegia in children through gummatous disease of the middle cerebral artery permitting thrombosis of that artery?

In adults there are two other ways in which syphilis leads to hemiplegia. There is epileptic hemiplegia associated with gummatous disease of the surface of the hemisphere, and there is hemiplegia caused by a gummatous growth in the motor tract. Possibly congenital syphilis in some cases causes hemiplegia in these two ways, as well as by thrombosis of a syphilitic vessel.

These remarks, which are certainly true of hemiplegia from syphilis in adults, shew that the seemingly definite expression "caused by" congenital syphilis is far from being a precise expression. The hemiplegia in the first results because there is softening of the corpus striatum; in the second it follows, and, as I believe, is caused by, a strong epileptic discharge; and in the third only is it caused by a syphilitic growth actually squeezing nerve-tissue.

I have had under my care recently a girl who had hemichorea. She had the dental malformation described by Mr. Hutchinson, and also remains of keratitis. I believe these to be certain signs of congenital syphilis. I see, however, no warrant for a decisive conclusion that the chorea was "caused by" the syphilis. The case was the sixty-second of about eighty investigated most elaborately by myself and Mr. Herman. It is the only one of the series in which there is evidence of congenital syphilis. It is true that in this girl's case there was no evidence of any other mode of causation. But this is not very unfrequently so in cases of chorea. So that if here I "eliminate other causes," we should conclude for congenital syphilis if we determined to come to a conclusion. But we are not bound to come to a conclusion without evidence. I do not, however, think there is even a probability of connection, for I have seen scores of cases of chorea essentially like the one alluded to but without any evidence of syphilis. In the pamphlet already mentioned I record a case of chorea occurring in a girl who was a member of a family in which there was clear evidence of syphilitic taint; she afterwards died of heart disease. I attended her mother

for rheumatic fever, and again for a fatal illness from heart disease. I also saw the child's brother for rheumatic fever. Here it was much more likely that the rheumatic taint was at the bottom of the mischief in the girl who had chorea.

Without any contradiction I may urge that nervous affections with congenital syphilis do not receive the attention they deserve. For the very reason that I am anxious to develop the subject, I have urged that in the absence of post-mortem examinations we can only arrive at probability. To speak decisively of symptoms as being *caused by* hereditary syphilis when there are no post-mortem examinations to refer to is only verbally decisive. It is verbally very definite to say "a square is round," but it is impossible to think so. He who speaks of hemiplegia and convulsion as "caused by congenital syphilis" is not only saying what he does not know, but what he cannot know. It may be said that from clinical evidence we conclude that lead-poisoning *causes* paralysis of the extensors of the forearm. But this is not a parallel case. The variety of paralysis is a rare one, and is scarcely ever found except in cases of lead-poisoning. If we saw a patient who was hemiplegic and the subject of lead-poisoning, we should not conclude that his hemiplegia was "caused by" lead-poisoning,* even if we could find no other explanation; we should leave the case unexplained rather.

So far as I have seen the hemiplegia and convulsion which occur with congenital syphilis do not differ from those symptoms as occurring in patients who present no evidence of that taint.

With regard to treatment, the person who concludes is in no better case for his conclusion than the other who waits. For no educated medical man would hesitate to treat syphilis whenever he found it, or, with reasonable exceptions, to treat local symptoms or symptoms implying local lesion, by anti-syphilitic remedies whenever he found them in a patient whose body presented signs of congenital syphilis.

Signs of Congenital Syphilis-Dementia.—Father died Insane.

The following notes were taken in the summer of 1872:—

Samuel L., aged 15, is the subject of congenital syphilis. The evidence supplied by his own person is (1) that he has that malforma-

* I do not mean that lead-poisoning may not lead *indirectly* to hemiplegia, for in some cases we find gout, chronic renal disease, and arterial degeneration. So, in a roundabout way, lead-poisoning may "cause" hemiplegia.

tion of the upper central incisor teeth which has been described by Mr. Hutchinson as characteristic of congenital syphilis. The lad's upper central incisors are small, and narrowed at their cutting edges; they are not notched.* (2) There are in the left eye remains of choroiditis; there are numerous pigmented patches. The left optic disc is atrophic (greyish). The cornea is clear.

The striking thing was that in the right eye there was nothing abnormal, although the left was much diseased. Both Mr. Hutchinson and Mr. Waren Tay agree in this. Obviously this non-symmetry in a disease so very "constitutional" as syphilis is of marked interest to students of disease of the nervous system. The choroid is the pia mater of the eye. This matter is so important that I must give a brief account of Mr. Hutchinson's opinion on the matter.

At first glance the one-sidedness of the condition appears to go against the diagnosis of congenital syphilis. Mr. Hutchinson, however, considers it to be the rule for choroiditis in connection with tertiary syphilis to be unsymmetrical, and this remark applies alike to that resulting from inherited and to that from acquired disease. It is not, however, he tells me, usual to find, as in this boy's case, that one eye is *quite* free from changes. He has, however, seen a few similar cases. The common condition is for one eye to be severely affected and the other only slightly. And he thinks that in some cases of choroido-retinitis in connection with hereditary syphilis in which the changes simulate those of retinitis pigmentosa the non-symmetry is a valuable point in diagnosis. Mr. Swanzy, of Dublin, has published an interesting example of this in the "Dublin Quarterly Journal," May, 1871. Mr. Swanzy there quotes a letter of Mr. Hutchinson's.

* The following extract from the pamphlet mentioned at the beginning of this article embodies some of Mr. Hutchinson's opinions :—"It is important to keep in mind the fact that this valuable test of the existence of a syphilitic taint in a family is to be found usually in but one of the children of that family. It is to be particularly observed that, although Mr. Hutchinson has described many dental peculiarities in children, he relies only—for a test of congenital syphilis—on a *certain* malformation of the two upper central incisors of the *permanent set*. Normally these teeth are chisel shaped, *i.e.*, *broad*er at their cutting edges than at their insertions into the gum. The malformation which Mr. Hutchinson has discovered to be a sign of congenital syphilis consists (First) in a reversal of the normal shape so far as this, that the two teeth above-named are *narrower* at their cutting edges than at their insertions into the gum. Hence they are then, as Mr. Dixon has observed, like 'screw-drivers.' (Second) The teeth are *often* notched. Hence such teeth are often called 'notched teeth.' It is well to add that Mr. Hutchinson attaches no special importance to 'bad teeth,' to 'irregular teeth,' &c., but, I repeat, to a particular kind of malformation of two of the permanent teeth."

Family History.—Two years before his marriage the father had a skin disease, and had a bad sore throat. He is dead; hence the vague history. Moreover, he died insane in Colney Hatch. The boy's mother, six or seven months after marriage, lost "all her hair" (no doubt an exaggerated expression); soon after marriage she suffered from a severe sore throat, which lasted seven or eight months; her tongue was very sore; she had a skin disease "like small boils or pimples."

The mother had seven children born alive. The following gives the results of all her pregnancies:—(1) Still-born. (2) Died at age of one month. (3). Samuel L., the subject of this report. (4) A child who now suffers from a skin disease. (5) Miscarriage. (6) Died at the age of five months; suffered from "snuffles," and had a skin disease. (7) A miscarriage. (8) A child who has a skin disease. (9) Died at the age of six-and-a-half months; "used to break out in the head," and suffered from snuffles. Nearly all the children, including our patient, suffered from a rash on the buttocks when infants.

It is important to note that our patient is the eldest living. This boy has not had keratitis. He may yet have it.* There is in my mind no doubt that this lad was the subject of congenital syphilis. The dental malformation, to say nothing of the choroiditis and of the family history, is, I think, decisive. A good many years ago I had the inestimable advantage of working with Mr. Hutchinson, and as a result of seeing many cases with him I was convinced that he was right in his assertion as to the diagnostic value of the malformation of the teeth he describes. So far as it goes, the family history supports the diagnosis founded on the dental malformation and the choroiditis.

Of course it is not said that congenital syphilis does not exist without either the dental malformation or the interstitial keratitis. On the contrary, it has been stated that these signs usually exist only in the eldest living of a syphilitic offspring. The younger children, no doubt, suffer in slighter degrees.

Normal Mental History.—The following facts as to the history were obtained after the lad's death by Mr. Mercier. The boy lived at a distance; he rarely attended, and thus the records were imperfect:—

* I remember once seeing in Mr. Hutchinson's practice a boy who had the dental malformation, and who was brought for amaurosis from optic atrophy; keratitis came on whilst he was being treated for the amaurosis.

As a child he was rather precocious ; he talked earlier than the other children ; mother cannot say at what age. Learnt to read and write early. During the greater part of his life he was by no means unintelligent ; he was quick and ready, had an excellent memory, was fond of reading ; wrote, his mother says, very good letters. He was careful of his money, but not miserly. Saved it up, and spent it upon clothes and other necessaries. For a short time he taught in a school, and the schoolmaster was satisfied with him. He left off because his family moved. He spoke well and sensibly on general topics (father-in-law instanced religion and the Abyssinian war). He never played much, but spent his spare time in reading and writing. He was obedient and submissive ; good-tempered, affectionate, not mischievous, not passionate. He was fond of music ; he was very religious.

He never did odd things, nor behave strangely. The only thing mentioned was that when he made a mistake in writing orders (which was his work) he always tore the page out instead of cancelling it, and of this he never could be cured. Now and then he would forget where he had laid things, and occasionally he would leave his medicine in the railway carriage.

Abnormal Mental Condition (1872).—He looked listless and sullen, and was evidently fatuous. I am obliged, however, since I only saw him as an out-patient, to rely for positive facts on what his mother told us. The following notes were taken for me by Mr. G. E. Herman. I incorporate with them some notes taken by Mr. Mercier :—

Some months before his actual mental failure a ship in the yard where he was at work caught fire, and this seems to have affected him a great deal. The other boys used to tease him, and say that he set it on fire, and he used to come home and ask his mother whether he could have done so. It is certain that he did not, and that he was some distance away when it happened. After this he became very much addicted to masturbation, then his memory began to fail ; he got dull and stupid, took no interest in things, but would sit by the fire and fall asleep. He became very sleepy, and would fall asleep under almost any circumstances. For several months before I saw him he had been in the habit of bringing home and hiding things that did not belong to him in the workshop ; he would hide the men's tools, and when they asked for them, "he looked so innocent, you'd never think it was

him.”* “We have told him we’d put him in prison, and such like, but it takes no effect, he doesn’t seem to care.” He did not take money, nor did he sell the things he carried away. “A short time before his father’s mental failure he was in the same way.” “He seemed so dull, and heavy for sleep.” “He is so dreadful forgetful; orders are always coming back.” “He writes so badly; he used to be a beautiful writer at one time.” “He drags along the streets just like an aged man.” His temper is very violent, “he is so spiteful; sometimes he seems so fond of his sisters, and other times would almost kill them.” He sleeps a great deal; often pulls off his clothes, and lies naked. His mother has seen a discharge on the sheet, “like dirty water,” which stiffens the sheet; a spot about as big as a five shilling piece. He has no sore.

He is fond of singing and music; has seemed rather more so lately. He soon ceased to attend. We afterwards learned that he died on October 13, 1874. The mother says that he died of some fever; she thinks scarlet fever, because his skin peeled when she wiped his face. There was some difficulty in getting the facts from the parents. For the last six months of his life he stammered, but no particulars of the stammering were to be obtained. He never lost power in either hand, never had a fit. A fortnight before his death he lost his speech altogether, but the mother is sure from his gestures that he retained his consciousness. There was no autopsy.

We are justified in concluding that this boy’s right choroid had at one time suffered from syphilis, and thus the *hypothesis* was, for the sake of treatment, warrantable that his *via mater*, the “brain’s choroid,” had suffered similarly,—had been the seat of a “*pia matritis*,” analogous to the choroiditis. My speculation is that there was local syphilitic disease, followed by general atrophy of the hemispheres. It is well known that extensive *local* damage (clot, softening, or tumour) in a hemisphere leads slowly to general wasting of that hemisphere.

Treatment did no good. But this did not negative the existence of syphilitic brain disease. Treatment is only likely to be useful during active syphilitic changes—as iritis, keratitis, ulcerations, nodes, &c. Besides in adults most syphilitic nervous symptoms are only indirectly owing to syphilis. To return to a previous example: the common form of hemiplegia in syphilis is owing directly to softening of the

* The words in inverted commas are his mother’s statements.

corpus striatum, to a condition just like that occurring from embolism. And when there is a syphilitic growth in the cerebral hemisphere, most active anti-syphilitic treatment in many cases that come late under our care only delays, does not prevent, a fatal issue. It would be as reasonable to expect to rid the pupil of lymph effused in an acute attack of iritis months before we saw the patient as to restore a brain damaged by syphilitic changes which were active months before. For general cerebral atrophy consecutive on local syphilitic disease we could do nothing.

If there were syphilitic disease of the brain it was in all probability of the connective tissue, and thus it was primarily non-nervous. In nearly all cases of nervous disease in which there are obvious post-mortem changes, it is plain that the changes are primarily non-nervous. Certainly all I know of syphilitic affections of the nervous system in adults is that they are diseases of the connective tissue entering into the composition of nervous organs or their arteries. I need not deny, however, that syphilis leads directly to changes beginning in nervous tissue; although I confess I do not see the evidence to warrant that conclusion. Let us again assume, as we did for the sake of treatment, provisionally, that syphilis did in some way cause this boy's mental illness. This is only the beginning of a difficulty. We cannot suppose that syphilitic or any other sort of disease of the cerebrum would *cause*, that is cause directly, such strange doings as those noted by Mr. Herman. It does not help us, to assume that the syphilis led to general cerebral atrophy. Atrophic tissue would "cause" nothing. Such doings are necessarily the result of action of parts of the brain which are *not* diseased. My own speculation is that the boy had by inheritance an imperfect brain, and that any sort of disease, syphilitic or otherwise, could have led easily to *loss* of the higher intellectual and emotional faculties, and indirectly from this "loss of control," or lack of inhibition, to increased play of the lower faculties. He had little higher faculty to lose.* The odd doings occurred from quasi

* We had a patient in the London Hospital, who, fatally ill from large cerebral hæmorrhage, was seen by Dr. Sutton and Mr. Lewis Mackenzie to twirl his moustache very elaborately every now and then. Except for this the man was to all appearance deeply comatose, and died a few hours later. It would be as reasonable to say that the clot of blood "caused" this moustache twirling, as to say that syphilitic disease or cerebral atrophy "caused" such symptoms as "hiding tools."

automatic action of centres left uncontrolled, the *disease* being of the highest or controlling centres.

The boy's father died insane in Colney Hatch Asylum, and this fact supports the inference above drawn. There were two factors, an imperfect brain by inheritance and disease.

The facts stated under the heading Normal Mental History do not invalidate the inference that the boy, besides receiving a syphilitic taint, received also an imperfect brain. For the facts show a precocious cleverness rather than intelligence, a style of ability quite consistent with a want of a high and robust intellect. Mere cleverness is no good sign of mental superiority; on the contrary, precocious cleverness in children is an evil sign. Cleverness is automatic intellect, and is comparable with instinct; the merely clever child does not go on developing, for he does not inherit much latent higher faculty to develop. Anstie gives us one sign of what is called the Insane Diathesis or Neurosis (I quote from Bucknill and Tuke), "Unexpected development of intense artistic feeling in children born of a naturally commonplace family." It is well known that idiots are often "clever" at music. In the Hospital Reports of the "*Lancet*," Feb., 1866, I mention the case of a speechless boy whose mental condition was very imperfect. His mother said, however, as evidence of the contrary, "But he has such a wonderful idea of music!" Since that report, I have seen many cases of grave mental inferiority in children who could sing and hum tunes correctly; cases of partial idiocy in fact. In an able paper in the "*Journal of Mental Science*," Oct., 1872, Dr. Ireland writes, "It is common enough for parents to found hopes upon their children having a good ear for music,* but this seems a gift common to all kinds and degrees of idiots."

The excessive masturbation is a fact of great clinical importance. Mr. Mercier says there was clear and abundant evidence of it. I do not, however, think it likely that it was a *cause* of the mental degradation in this case. I think it was rather the consequence. In all diseases affecting the mind, there is a reduction, not only to a lower intellectual level, but to a lower level of feeling, and in slight degrees

* "On retrouve les traces de cette persistance partielle de l'intelligence jusque dans les formes les plus avancées des dégénérescences héréditaires. Il n'est pas rare, au milieu de l'anéantissement des facultés psychiques qui caractérisent l'idiotie héréditaire, de voir survivre une faculté intellectuelle. Ces êtres dégénérés possèdent pour la musique, le calcul, le dessin, la poésie, etc., une aptitude instinctive, native, qui paraît d'autant plus extraordinaire que le contraste la fait briller d'avantage."— *La Folie Héréditaire* par le Dr. Legrand du Saulle.

there is seen to be only an apparent intensification of the inferior parts of the natural disposition. The vulgar man talks and acts offensively, the spiteful man becomes actively malicious, and the sensual man is openly indecent. I believe, then that, as a rule, insane patients masturbate as a consequence of disease of the brain, and from the same cause that many of them become peevish and greedy. There are some cases in which there is a strong warrant for the assumption that masturbation primarily causes grave mental disease, but in some of the best marked cases I have seen there has been inherited predisposition to mental disease. Obviously mental degradation, however begun, would be helped on by masturbation. The mental disease introduces a new or secondary cause of mental depravation.

I would here remark that I do not believe masturbation causes epilepsy, or chorea, or any such symptoms, any more than I should believe it could cause paralysis of the portio dura nerve. It is sometimes replied to such a statement as this, that it is a "question of fact." It is not a question of fact with me, because I admit the fact, that is, I admit that many epileptics masturbate. All I deny is the inference that masturbation has caused their epilepsy. That it is one of the gravest factors in producing mental deterioration is no proof that it leads to an entirely different class of symptoms, any more than the fact that lead-poisoning causes paralysis of the extensors of the forearms is proof that it causes hemiplegia.

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